

Persistence and effects of sinus rhythm after Fontan procedure for tricuspid atresia

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SUMMARY Four patients who had had a Fontan type of procedure for tricuspid atresia 23, 6, 9, and 11 months previously were investigated by ambulatory electrocardiographic recording and simultaneous recording of the jugular venous pressure and echocardiogram of the conduit or pulmonary valve. All had been considerably improved by the operation. In 1 patient episodes of supraventricular tachycardia were recorded but no rhythm disturbance was detected in the other 3. Pulmonary blood flow was shown to be pulsatile and atrial systole is an important factor in this. The conduit valve showed delayed opening and slow closure suggesting that its presence in the pulmonary circuit may be unnecessary.

In 1971 Fontan and Baudet reported 3 patients with tricuspid atresia in whom they redirected the entire systemic venous return to the pulmonary circulation by attaching the superior vena cava to the right pulmonary artery and the right atrium, through an aortic homograft valve, to the pulmonary artery bifurcation and hence the left lung. A homograft pulmonary valve was inserted into the inferior vena cava, the atrial septal defect closed, and the main pulmonary artery ligated.

With more experience of the technique (Ross and Somerville, 1973; Henry *et al.*, 1974; Somerville and Ross, 1975; Stanford *et al.*, 1975; Walker *et al.*, 1975), modifications have been introduced. In some cases the right atrium has been connected to the outlet chamber (Somerville and Ross, 1975) and there is debate regarding the need for caval valves (Fontan *et al.*, 1977; Yacoub, 1977). The technique has also been extended to other forms of the hypoplastic right heart syndrome (Yacoub *et al.*, 1975; Sharratt *et al.*, 1976).

In all variants of the operation atrial systole appears to be necessary for a good result (Ross and Somerville, 1973; F. Fontan, 1977, personal communication). The present study was performed to investigate the role that atrial systole plays in pulmonary blood flow and to see if there are any indications that sinus rhythm may not be maintained after this procedure.

Subjects and methods

Four patients with tricuspid atresia, who had successfully undergone the Fontan operation, were studied.

Echocardiograms and the externally derived jugular venous pulse wave (JVP) were recorded on a Cambridge strip-chart recorder. Echocardiography was performed using a Smith Klein Ekoline 20 and a 2.25 MHz transducer focused at 5 cm. The transducer position varied according to the site of the conduit or pulmonary valve.

Ambulatory electrocardiographic recordings were made on an Oxford Medilog dynamic cardiogram recorder and these were analysed on a Reynolds Pathfinder electrocardiograph analyser.

CASE 1

This patient was born in 1951. At the age of 6 years a left Blalock-Taussig shunt was performed for increasing dyspnoea and cyanosis. In 1965 he had his first episode of palpitation and these occurred occasionally from then on. The electrocardiogram showed sinus rhythm with a PR interval of 0.09 s (Fig. 1A) suggesting the presence of the Lown-Ganong-Levine syndrome. Periods of high junctional rhythm (coronary sinus rhythm) (Fig. 1B) and a tachycardia of 130/min arising from this same focus (Fig. 1C) were also recorded.

In 1973 repeat cardiac catheterisation disclosed atrial situs solitus, univentricular heart with anterior outlet chamber, discordant arterial connections with

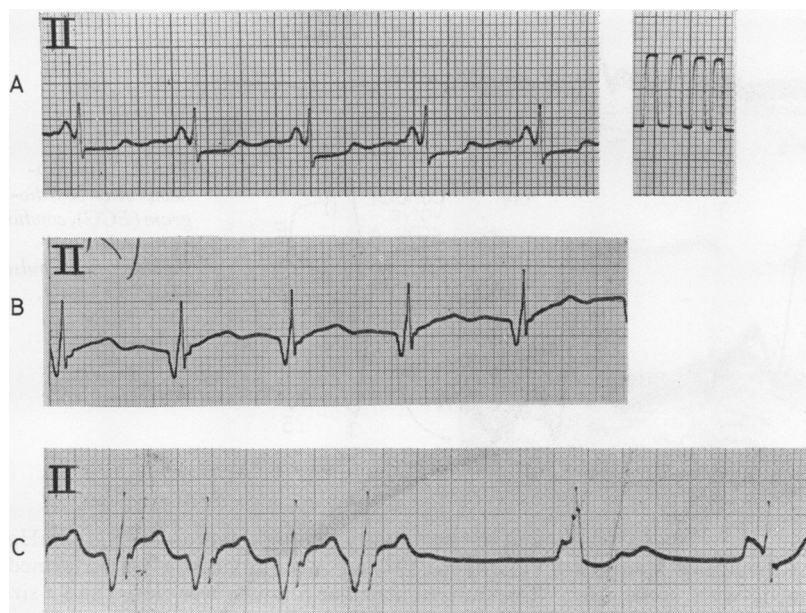


Fig. 1 Preoperative electrocardiograms in case 1.

pulmonary outflow tract obstruction, and atresia of the right atrioventricular valve and a secundum atrial septal defect. The Blalock-Taussig shunt was virtually functionless. The pulmonary artery pressure was 12/5 mmHg. On 16 April 1975 a Fontan procedure was performed. The main pulmonary artery was ligated and a size 21 mm homograft valve conduit inserted between the right atrial appendage and the right pulmonary artery. The atrial septal defect was closed by direct suture. No valves were placed in the inferior or superior vena cava.

He was asymptomatic 32 months after the operation and leading a full and active life as an electrical engineer. The jugular venous pulse had an A wave visible to 6 cm above the sternal angle but there was no hepatomegaly or oedema.

CASE 2

This boy was born in 1969. At the age of 10 months a left Blalock-Taussig anastomosis was performed with some improvement. By 1977, however, he had deteriorated to such an extent that he was almost confined to a wheel-chair. The diagnosis was dextrocardia, visceral and atrial situs solitus, and double outlet univentricular heart without a rudimentary chamber; the aorta was anterior and slightly to the right of the pulmonary artery; there was pulmonary outflow tract obstruction, and atresia of the right atrioventricular valve with a secundum atrial septal defect and a non-functioning left Blalock-Taussig anastomosis. At operation on 24 August 1977, the main pulmonary artery was ligated and a size 18 mm

antibiotic sterilised homograft valve was inserted between the right atrial appendage and the right pulmonary artery. The atrial septal defect was closed by a pericardial patch and no caval valves were inserted. Six months later he was asymptomatic and beginning to take part in physical education at school. There was a 4 cm A wave in the jugular venous pulse but no hepatomegaly or peripheral oedema.

CASE 3

This girl was born in 1965. In 1967 a right Blalock-Taussig anastomosis was performed. She remained fairly well with moderately reduced effort tolerance. Reinvestigation indicated atrial situs solitus, univentricular heart with an anterior outlet chamber, and normally connected great arteries. There was pulmonary outflow tract obstruction, atresia of the right atrioventricular valve, and a secundum atrial septal defect. The right Blalock-Taussig shunt was not functioning. On 20 April 1977 a Fontan procedure was performed. The main pulmonary artery was ligated and a size 18 mm antibiotic sterilised homograft valve was inserted between the right atrial appendage and the right pulmonary artery. The atrial septal defect was closed with a pericardial patch and the old Blalock-Taussig anastomosis was ligated. No caval valves were inserted. Eleven months later she was asymptomatic and able to take part in physical education at school. She had a 5 cm A wave in the jugular venous pulse but no hepatomegaly or peripheral oedema.

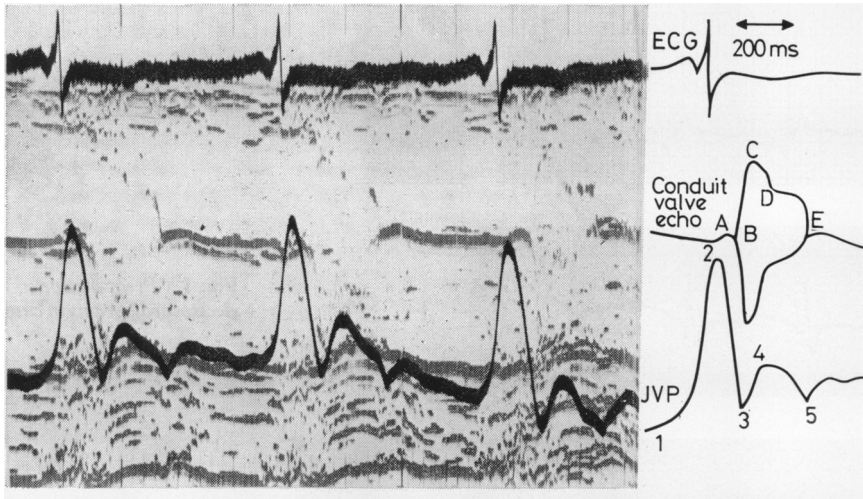


Fig. 2 Simultaneous electrocardiogram (ECG), conduit valve echo, and jugular venous pulse (JVP) in case 1. See text for description.

CASE 4

This boy was born in 1963. At the age of 5 years a Waterston shunt was performed, with some improvement, but he became progressively more cyanosed and effort tolerance declined. At repeat cardiac catheterisation in 1973 atrial situs solitus, univentricular heart with anterior outlet chamber, normally connected great vessels with pulmonary outflow tract obstruction, atresia of the right atrio-ventricular valve, and a secundum atrial septal defect were confirmed. The Waterston anastomosis was still functioning. Pulmonary vascular resistance was 14 per cent of the systemic vascular resistance

and the pulmonary artery pressure was 18/4 mmHg. On 29 June 1977 a Fontan procedure was performed. The bulboventricular foramen was closed and a size 18 mm antibiotic sterilised homograft valve was inserted between the right atrial appendage and the outlet chamber. The atrial septum was closed with a pericardial patch and the Waterston shunt was closed. No caval valves were inserted. Recovery was uneventful and 9 months after the operation he was asymptomatic and played football, basket ball, cricket, and golf. There was a 6 cm A wave in the jugular venous pulse with no hepatomegaly or oedema.

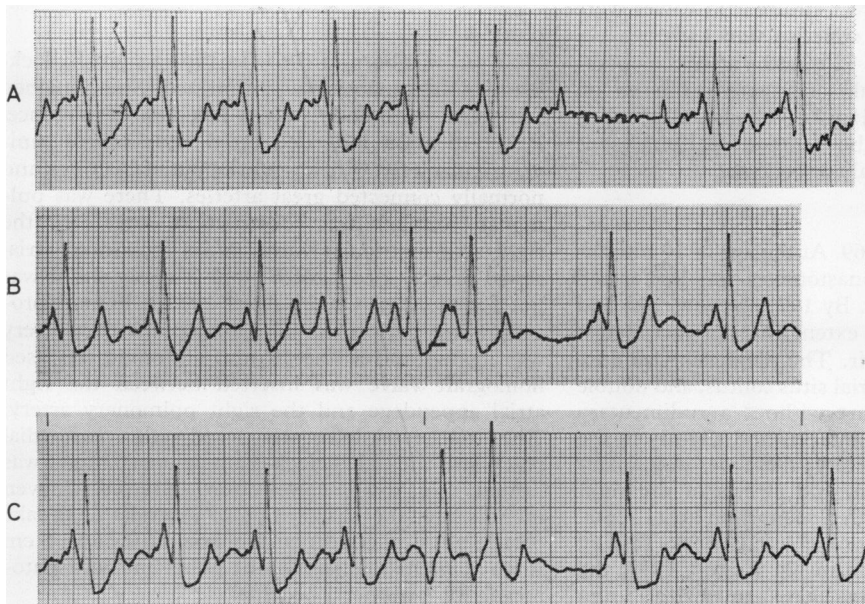


Fig. 3 Extracts from ambulatory electrocardiogram in case 1.

Results

CASE 1

Echocardiography (Fig. 2)

Opening of the conduit valve (B) begins 80 ms after the peak of the A wave of the jugular venous pulse waveform (2). It is preceded by a small anterior movement (A). The X descent begins before the valve opens and continues to decline sharply until the valve achieves its maximal opening (C). The cusps then become more closely apposed (D) and there is a small rise in the jugular venous pulse (4). The valve then closes (E) and there is a brief dip in the jugular venous pulse (5). The valve is open for 250 ms.

Ambulant electrocardiogram

The sinus rate varied from 70/min (during sleep) to 125/min. The only conduction disorder recorded is shown in Fig. 3A. Two types of atrial extrasystoles were recorded, one with an upright P wave, and one with an inverted P wave similar to the preoperative abnormal rhythm. Only 4 of the first type were recorded, 1 in isolation and 3 in succession (Fig. 3B). Twelve of the second type were recorded, 5 in isolation, 2 periods in pairs (Fig. 3C) and 1 episode of 3 in succession. Ninety-six ventricular extrasystoles were recorded, all as single beats. The tape time was 24 hours.

CASE 2

Echocardiography (Fig. 4)

The echo of the conduit valve is incomplete but it is seen in its fully open position and during its closing

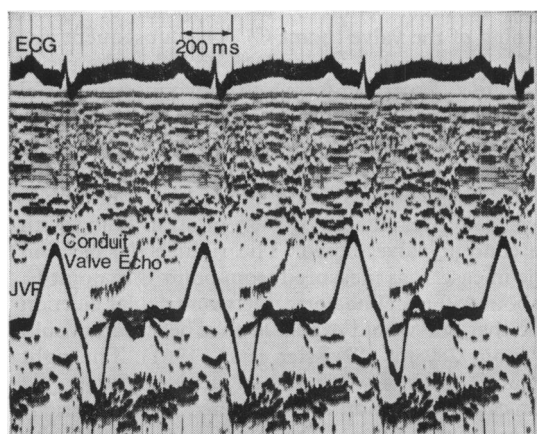


Fig. 4 Simultaneous electrocardiogram (ECG), conduit valve echo, and jugular venous pulse (JVP) in case 2.

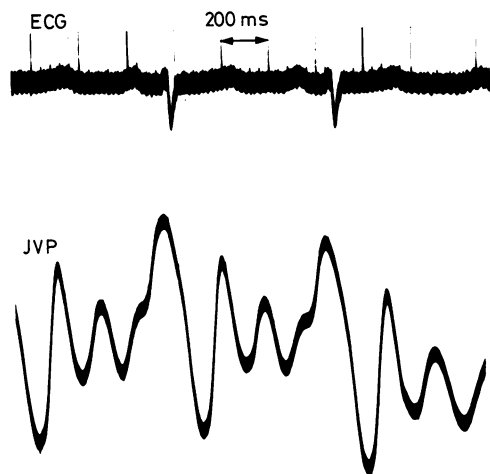


Fig. 5 Electrocardiogram (ECG) and jugular venous pulse (JVP) in case 3.

movement, which is more gradual than in case 1. The jugular venous pulse is very similar to that described for case 1.

Ambulant electrocardiogram

The heart rate varied from 65/min (during sleep) to 110/min. There were 4 atrial extrasystoles and 1 ventricular extrasystole recorded during the 22-hour recording period.

CASE 3

Echocardiography

It was not possible to record the conduit valve and

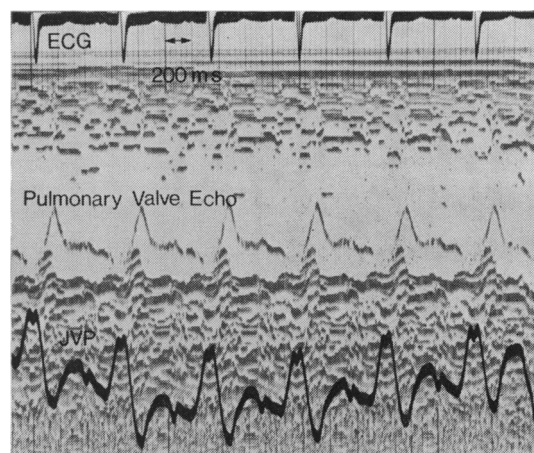


Fig. 6 Simultaneous electrocardiogram (ECG), pulmonary valve echo, and jugular venous pulse (JVP) in case 4.

no pulmonary valve was present in the pulmonary circulation. Jugular venous pulse (Fig. 5) was similar to those recorded in the other patients.

Ambulant electrocardiogram

The heart rate varied from 83/min (during sleep) to 150/min. Five ventricular extrasystoles and 3 atrial extrasystoles were recorded. The tape time was 24 hours. Tape analysis was complicated by a large P wave which could not be reduced by changing the electrode position.

CASE 4

Echocardiography

The conduit valve could not be recorded but the patient's own pulmonary valve was recorded. It is seen to follow a similar pattern to that of the conduit valve in case 1 (Fig. 6). Opening begins 40 ms after the peak of the A wave of the jugular venous waveform, the valve starts to open and its point of maximum opening coincides with the nadir of the dip in the venous wave. There is then partial closure of the cusp. The point of final closure is not accurately defined.

Ambulant electrocardiogram

The heart rate in sinus rhythm varied between 72/min (during sleep) and 150/min. Eleven isolated atrial extrasystoles were recorded (Fig. 7A), a 20-second burst of atrial tachycardia with a rate of 146/min (Fig. 7B) and a 30-second burst with a rate of 200/min (Fig. 7C). The first episode was asymptomatic while during the second episode the patient experienced severe cramp in a leg though it is not

clear whether this was the initiating event or occurred during the paroxysm. The tape time was 22 hours.

Discussion

Severe vascular changes may occur in the right lung after a superior vena cava pulmonary artery anastomosis (Achtem et al., 1969; McFaul et al., 1976). This appears to be related to sluggish blood flow (Laks et al., 1977). It, therefore, seems important after the Fontan procedure for tricuspid atresia that atrial systole is maintained if it is true that atrial systole contributes significantly to the rate of pulmonary blood flow. This certainly appears to be the case as the loss of atrial systole, for example when atrial fibrillation occurs, results in systemic congestion which resolves after the restoration of sinus rhythm by DC cardioversion (F. Fontan, 1977, personal communication). The present investigation was undertaken in an attempt to delineate the contribution of atrial systole to pulmonary blood flow and to assess the likelihood that sinus rhythm might not be maintained in the 4 patients studied.

Echocardiograms of the conduit valve in cases 1 and 2 and of the pulmonary valve in case 4 certainly showed that pulmonary blood flow was pulsatile and that atrial systole was an important factor in this. The jugular venous waveform appeared to have a very similar pattern in all 4 patients. We believe that the sequence of events, shown in Fig. 2, is right atrial isovolumic contraction (point 1 to point B) which moves the closed valve slightly forward (point A). The valve appears to have some inertia which has to be overcome, the jugular venous pulse declining before the valve opens. With opening of the valve there is a sharp fall in the venous waveform, maximal (point 3) at the point of widest opening of the valve (point C). The subsequent rise in the venous pulse (point 4) is probably the result of venous return, entering the atrium with atrial relaxation. At this point the valve is still open but less widely so (point D). There follows a slow fall in the venous pulse then to point 5 at which time the valve closes (point E) and the venous pulse then rises again presumably because of atrial filling with the conduit valve closed. The right atrial ejection time in case 1, as measured from point B to point E, is about 250 ms. The normal left ventricular ejection time that one would expect at that heart rate would be about 280 ms (Weissler et al., 1961). The right atrium is therefore acting as an efficient pump.

In case 4 the conduit was attached to the ventricular outlet chamber. The pulmonary valve echo would therefore be expected to show the effects of outlet chamber systole. However the movement of

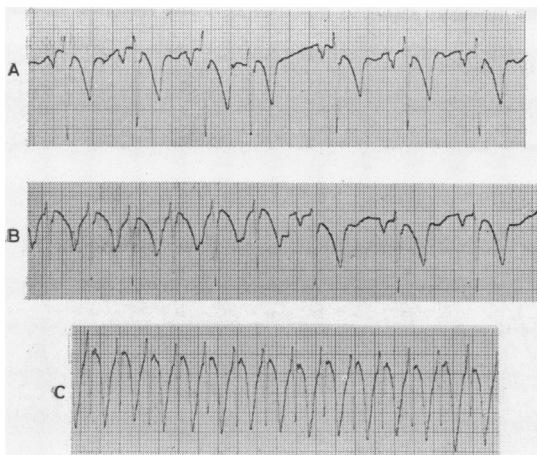


Fig. 7 Extracts from ambulatory electrocardiogram in case 4.

the pulmonary valve (Fig. 7) is similar to that of the conduit valve in the other patients, and there is no additional opening movement that could be attributed to contraction of the outlet chamber. The outlet chamber was known to be small.

It is difficult to reach firm conclusions from the results of the ambulant electrocardiographic monitoring because of the lack of control data. The minimum sinus rates of cases 1, 2, 3, and 4 were 70, 65, 83, and 72 beats per minute, respectively. These are high when compared with the normal population studied by Brodsky *et al.* (1977). There the minimum sinus rates were between 37 and 65 beats per minute but the population was older at 23 to 27 years. In cases 2 and 4 no untoward events were recorded. The atrial tachycardia in case 4 must raise a question as to the long-term outlook for the maintenance of atrial systole in him. Case 1, despite the preoperative occurrence of atrial tachyarrhythmias, had only 1 burst of 3 atrial extrasystoles in a row

but he did have 96 ventricular extrasystoles over the 24-hour period, which is probably excessive (Brodsky *et al.*, 1977). It is of interest that he was the oldest patient operated on and dysfunction of the primitive ventricle may occur if it has to handle a large volume for a long time (F. Fontan, 1977, personal communication). This may be relevant to the occurrence of the extrasystoles in this patient.

There is no doubt that there has been dramatic symptomatic improvement in these patients in the short time that they have been followed (23, 6, 9, and 11 months) and there is not the hazard of a right-to-left intracardiac shunt which remains in other forms of palliation for tricuspid atresia.

Our results encourage us to continue using this procedure though the arrhythmias in case 4 give cause for concern. It is our impression that caval valves do not improve the haemodynamics and indeed in view of the apparent opening inertia of the conduit valve it could be questioned whether any valves are required at all. When the outlet chamber is used, we believe that a valve should be inserted between the right atrial appendage and the outlet chamber. This conclusion is based upon our experience with 2 patients, not fully reported here. In one of these patients a 'dacron' tube was used and the patient died after 2 years, with systemic venous congestion. At necropsy, however, a fenestrated mitral valve cusp was found and this must have produced significant mitral regurgitation. In the second patient, recently operated upon, a valved conduit was used between the right atrial appendage and a large outlet chamber. The postoperative course was uneventful and the echocardiogram of the pulmonary valve (Fig. 8) shows no opening inertia, the rapid opening movement coinciding with the onset of the upstroke of the A wave of the jugular venous pulse, and the valve remains fully open during what appears to be an X descent and V wave. The valve closes at the peak of the V wave. There is thus, in this patient, clear evidence of outlet chamber systole. However case 4 in this series indicates that a small outlet chamber, after it has been opened and a conduit inserted, has little or no mechanical activity.

We thank Mr M. Sturridge for permission to study case 1 on whom he performed the operation.

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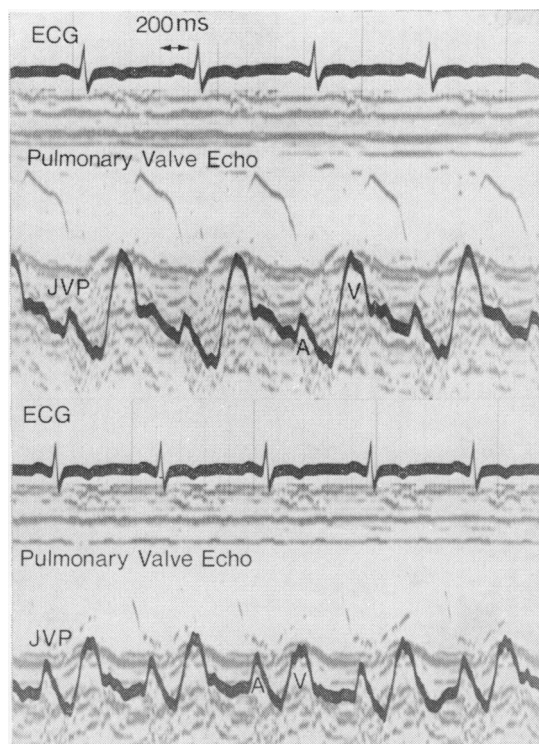


Fig. 8 Simultaneous electrocardiogram (ECG), jugular venous pulse (JVP), and echocardiogram of the pulmonary valve in the presence of valved conduit attached to a large outlet chamber. The top tracing shows the opening movement of the pulmonary valve; the bottom tracing shows the valve in its open position.

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